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Distribution of causes and outcomes of pulmonary hypertensions in a tertiary pediatric hospital

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Objectives: To describe the distribution of causes of pediatric pulmonary hypertension (PH) based on the experience of a pediatric PH department in a tertiary hospital with pluridisciplinary approach including rare diseases.

Background: The causes of PH have been well characterized in adult and pediatric large registries but the type of PH seen in expert centers has been modified by improved screening and diagnosis strategies.

Methods: Between 2008 and 2010, all PH patients referred to our PH department were included in a PH data base and classified according to the Dana Point clinical classification. Clinical, hemodynamic features, and outcomes of the first 212 cases are described.

Results: The median age at diagnosis of PH was 2.4 years (range 0.4-15); 56% were girls. At diagnosis, 40% of the patients were in functional class III or IV. Mean pulmonary artery pressure at diagnosis was 54.6±16.6 mmHg, PVR was 16±7 Wood Unit and cardiac index 3.2±0.8 l/min/m^2. Of 212 cases, 160 (75%) were in group 1 pulmonary arterial hypertension (PAH): idiopathic or heritable in 38 children and, associated with congenital heart disease (CHD) in 122 (57.5%). Six children were in group 2 PH (left heart disease), 30 (14%) were in group 3 PH (8 diaphragmatic hernias, 13 bronchopulmonary dysplasias, 5 interstitial lung diseases, 4 other causes), no children of our series were in group 4 PH, and finally 16 out of the 122 (7.5%) were included in group 5. At last follow-up, 93% of the PAH patients had received PAH specific drugs, and overall 85% of patients remained in a stable condition. Survival after three years follow-up was 94%,

Conclusion: The spectrum of causes, diagnostic challenges and gender distribution of pediatric PH in our national pediatric PH expert center are different to that in adults but also to that described in large pediatric registries. Still, clinical classification of PH appears appropriate in pediatrics. Our study also highlights current management and shows an improved prognosis with more aggressive therapeutic options

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A new anatomic approach to the ventricular septal defect in interruption of the aortic arch

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Objectives: To analyze the anatomy of the ventricular septal defect (VSD) in heart specimens with interruption of the aortic arch (IAA), in order to confirm the hypothesis of different embryologic mechanisms for the different anatomic types of IAA.

Material and methods: We examined 27 hearts from the anatomic collection of the French Reference Center for Complex Congenital Heart Defects with IAA, concordant atrioventricular and ventriculoarterial connections, and 2 distinct great arteries. Hearts were classified according to Celoria and Patton: type A, interruption distal to the distal subclavian artery (A), type B, between the distal subclavian and the carotid artery (B), type C, between the 2 carotid arteries (C). We focused on the anatomy of the VSD viewed from the right ventricular side.

Results: There were 10 A, 17 B, no C. One A (with aortopulmonary window) and 1 B had no VSD. The VSD was conoventricular, located between the 2 limbs of the septal band (LSB), in 4/9 A and 16/16 B (p=0.005), with posterior deviation of the outlet septum. In A, the VSD was conoventricular in 4 with muscular rims in 2 and fibrous extension of the posterior LSB in 2, muscular in 3, membranous in 2. In B, the VSD had entirely muscular rims in 4, fibrous extension of the posterior LSB in 9, and was juxta-arterial in 3; there was no fibrous continuity between the tricuspid and aortic valve.

Conclusion: The VSD in IAA type B is always conoventricular, with posterior deviation of the outlet septum, but without any fibrous tricuspid-aortic continuity. The VSD in IAA type A can be of any type. This reinforces the hypothesis of different pathogenic mechanisms responsible for the 2 types of IAA, and the inclusion of IAA type B in the group of conotruncal defects. The absence of fibrous tricuspid-aortic continuity indicates that the fibrous extension of the posterior LSB in some hearts may be due to the deviation of the outlet septum, due to an excessive rotation of the outflow tract.

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Longitudinal left ventricular strain impairment in type 1 diabetic children: a prospective 2D speckle strain imaging study

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Background: The relation between type 1 diabetes and cardiac structure and function in children is poorly documented. We used 2D speckle strain imaging to investigate whether children and adolescents with type 1 diabetes have early echocardiographic signs of subclinical cardiac dysfunction and whether state of metabolic control and diabetes duration are of influence.

Methods: Standard 2D echocardiography, mitral TDI and 2D speckle strain imaging were prospectively performed in type 1 diabetic children and compared them to age and sex- matched healthy control subjects. Standard echocardiographic indices of global systolic and diastolic function, early peak diastolic mitral velocity (Ea), longitudinal strain (LS), radial strain (RS) and circumferential strain (CS) were investigated.

Results: Overall 49 consecutive type 1 diabetic children (age: 12.3 [6-18] years; BMI: 19±2.8; males: 20) were compared to 49 control subjects. There was no difference between two groups for mean arterial blood pressure, heart rate, left ventricular ejection fraction, left ventricular mass, conventional diastolic mitral Doppler parameters (E, E/A, MTD) and Ea. A significant decrease of LS was identified in type 1 diabetic children while RS and CS did not differ. LS was positively correlated with HbA_{1c} (r: 0,28 ; p<0,001) and with diabetes duration (r: 0,20; p<0,001).

Conclusion: We demonstrated that left ventricular longitudinal myocardial strain is decreased in young patients with uncomplicated type 1 diabetes. Metabolic control may be the main risk factor for these myocardial changes. This finding might be considered a very early preclinical alteration potentially related to subsequent development of diabetic cardiomyopathy.

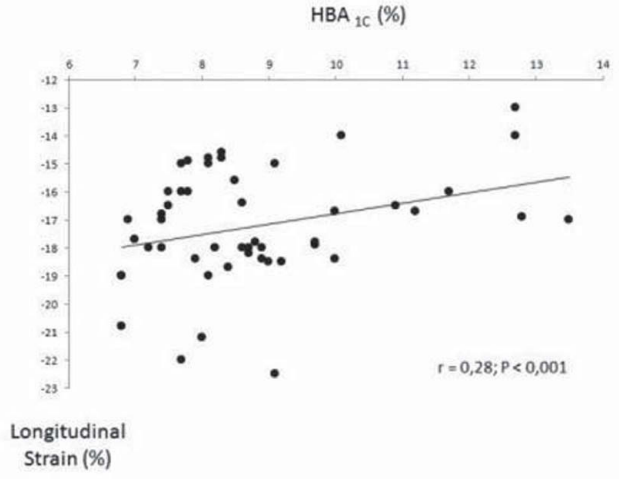

Figure: Correlation between Longitudinal Strain and HbA1C